

Pectus Deformities

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PECTUS DEFORMITIES are developmental asymmetries of the chondrosternal area in which the sternal plate is either depressed or elevated. Pectus excavatum (funnel chest) and pectus carinatum (pigeon breast) are the two main types (Figure 1) but sometimes the features of both occur in one person. Defects of the thoracic wall, such as bifid sternum and traumatic deformities, are not within the scope of this report.

Terms such as *trichterbrust*, *chonechondrosternon*, *pectus recurvatum*, *thorax en entonnoir* and *koilosternia* have been coined for depression defects, and *pectus gallinatum* for pigeon breast.

Pectus excavatum is characterized by a broad funnel-like depression of the lower anterior thorax, including the sternum and costal cartilages. The anterior-posterior diameter of the chest is shortened. The posterior deflection of the sternum usually begins near the third chondrosternal junction and is deepest at about the level of the tenth thoracic vertebra. The patient characteristically has kyphosis, stoop-shoulders, a "pot-belly" and other musculoskeletal irregularities (Figure 2).

In *pectus carinatum* the sternum is deflected anteriorly, forming a ridge, the xiphoid process being most prominent. A narrowing of the lower chest cage exists in the carinatum deformity (Figure 3).

History

Medical records of Johann Bauhinus¹ in 1594 contain the first clinical description of a patient with this problem, with the suggestion that a foreshortened diaphragm caused the sternal depression. For more than three hundred years, exercises and braces were the only therapeutic procedures known or employed.

The era of surgery was opened by Meyer's procedure¹³ (1911) in which he excised the second and third costal cartilages on the right side, bringing about slight lessening of the deformity. Two years later Sauerbruch¹⁷ excised the left fifth to tenth costal cartilages with adjacent one-fourth of the sternum, thereby providing fair relief of deformity and symptoms. Up to World War II only 27 patients had been operated upon. Six of them—all more than 16 years of age—died. In only half of the cases was the result of the operation a

- The surgical correction of a deformity of the anterior chest wall is becoming more frequent as knowledge regarding causes increases and as better results are achieved.

The present etiological concept of an excessive costal cartilage growth has prompted surgeons to perform total excision of the involved costal cartilages. The operation used in 27 patients included in the present study incorporated this principle and, in addition, the use of a temporary pin support until the flaccid chest wall becomes firm enough to resist a flail motion.

success. The procedures used in the 27 cases consisted of one or more of the following: Division, wedging or excision of costal cartilages, and "T" sternotomy, transverse osteotomy, sternectomy or inversion of sternum. In none of the procedures were traction devices, transfixion devices or internal splinting used for fixation.

In 1939, Brown² presented his surgical approach based on the "substernal ligament" hypothesis, in which he merely transected this ligament and removed the xiphoid process. This procedure is no longer recommended. The main objections are that it does not fully restore normal contour, the technical risk is great and there is likelihood of a recurrence of the deformity.^{2,9,11,15}

Improved techniques for anesthesia have encouraged a more radical and extensive correction of

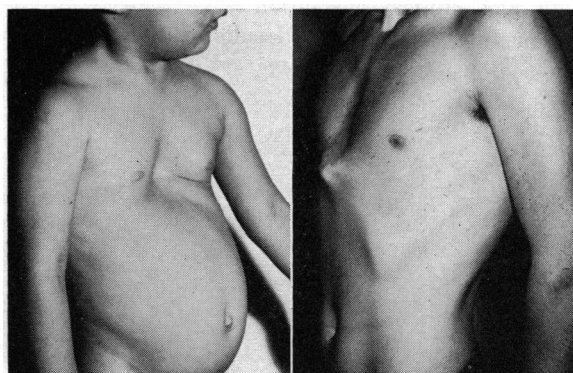


Figure 1.—The characteristic sternal depression of pectus excavatum (left) with stoop shoulders and pot belly. There is also an anteroposterior foreshortening of the thoracic cage. Pectus carinatum deformity (right) usually has some degree of lateral narrowing of the thoracic cage beside the sternal protrusion with adjacent wall depression.

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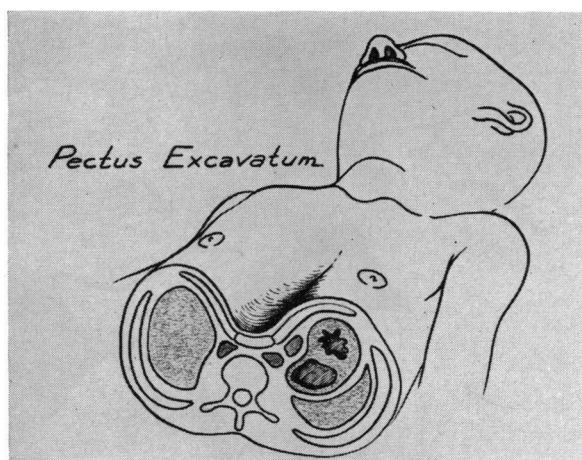


Figure 2.—The severe excavatum depression produces a marked shift of mediastinal structures to the left causing cardiac displacement and reduction of pulmonary function.

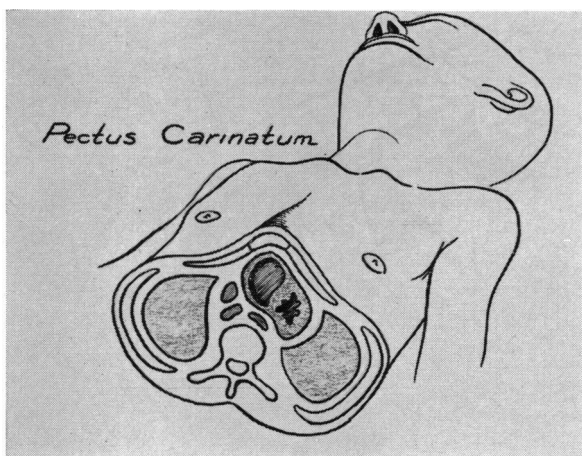


Figure 3.—Fixation of the deformed rib cage reduces the vital capacity even though organs may not be displaced noticeably.

pectus deformities and in more and more cases successful reconstruction is being achieved. At the same time, the mortality rate has been reduced to that of thoracotomy.

Incidence

Six of 10,000 babies born have pectus deformities. The incidence is four times as high in males as in females. The proportion of excavatum to carinatum anomalies is also four to one. The familial occurrence is noteworthy: One observer reported 11 members in four generations of one family had the deformity, eight in another and seven in three generations of a third family have been reported.¹⁴ In one instance twins were involved, one having a pectus excavatum and the other carinatum pectus. The trait is believed recessive, but in some circumstances it appears to be dominant.¹⁶

Etiology

Many theories as to causes for the deformity have been proposed. The four most significant hypotheses are:

1. That there is a shortened anterior-posterior fibromuscular structure behind the xiphoid process—perhaps the diaphragm or the central tendon or the anterior diaphragm muscle or the substernal ligament.^{2,8} However, in deformity of the excavatum type, the greatest depression of the sternum is usually located at or above the level of the tenth thoracic vertebra, which is above the origin of the diaphragmatic muscle. Also the sternum may be depressed into the thoracic paravertebral gutter, a site not related to substernal ligamentous attachments.

2. That there is an imbalance in the pectoral muscular innervation and function, with pigeon breast occurring when there are spastic or overstimulated muscles, and funnel chest where weak muscles permit the stronger diaphragm to “overpower” the pectoral groups.⁸

3. That there is abnormal attachment of the pectoral muscles in such a way as to affect the balance of muscle pull against the diaphragm. It is thought an insertion of the pectoral muscle at the sternum causes sternal protrusion and a lateral insertion causes sternal depression.⁸

4. That an overgrowth of the costal cartilages (or ribs) causes buckling in or out of the cartilages and sternal plate.^{7,10} The incidence of pectus deformities in Marfan's syndrome in which overgrowth of the long bones is a cardinal defect, tends to support this theory. We consider it the most plausible with regard to pectus deformities.

Less commonly ascribed as causes are intra-uterine pressure of chin, elbow or knee. In cobblers, constant pressure of the shoe-last against the chest is thought to be a cause.

Anatomical dissections and surgical evidence do not constantly support the hypotheses in Paragraphs 2 and 3 above. There is no proven cause.

Associated Anomalies

As is true with most developmental anomalies, pectus deformities are very frequently accompanied by other congenital defects. Of 25 patients observed by us, 12 had associated musculoskeletal or neurological problems. Some observers^{15,16} have reported associated anomalies in 30 per cent of cases.

Clinical Observations

The disturbances of function of cardiovascular, respiratory and digestive systems are difficult to measure, for the patient usually adapts to the deformity. Organs adjacent to the sternum are mobile and flexible. There is wide variation in the degree

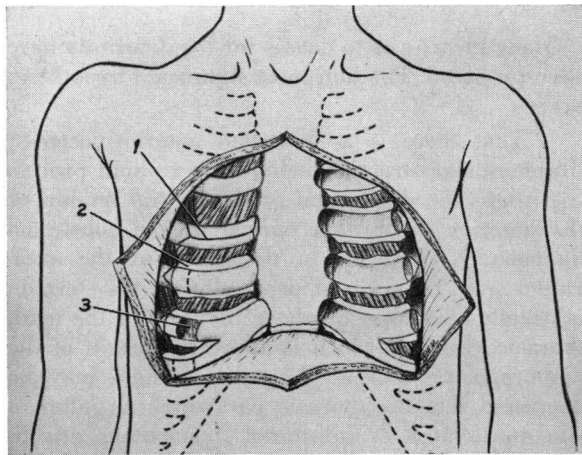


Figure 4.—Composite view of operative field with reflected flaps of inverted "Y" incision. (1) Perichondrium incision, (2) reflected, (3) segment excised. The left side of thoracic wall demonstrates the reflected perichondrium of the excised cartilages.

of deformity. In most cases the asymmetry is slight, in a few moderate. Extreme deformity is rare.

Cardiovascular problems are associated with great displacement of the sternum. The symptoms consist of dyspnea, palpitation, precordial pain, easy fatigue and fainting spells. Sometimes congestive failure occurs. Displacement of the heart to the left depresses the inflow and outflow volumes by compression or torsion on the great vessels.

Electrocardiographic and vectorcardiographic changes have been noted.^{19,20} Arrhythmia, ventricular tachycardia, P-wave changes, elevation of ST segment and T wave inversion have been demonstrated.^{4,5,20}

Exertional dyspnea is a common concomitant, and also, in extreme deformity, shortness of breath. There is a tendency to frequent recurrence and greater severity of infectious diseases of the respiratory tract because of a diminished ability to cough effectively. About a third of the patients we observed spoke of this tendency.

The respiratory excursions of the diaphragm are decidedly impaired and the expansion of the rib interspaces is decreased. Vital capacity is diminished.⁸

Parents are distressed by the deformity in infancy but the child, barring another anomaly incompatible with life, grows at normal rate. His distress begins as he becomes a physical curiosity to his playmates and it often increases as he approaches adolescence. Often as a teen-ager he shies away from locker rooms and from sports in which the torso is exposed.

Surgical Treatment

Minor deformities can be left untreated, for pectoral muscle development alone may suffice. If the deformity is great enough to consider braces,

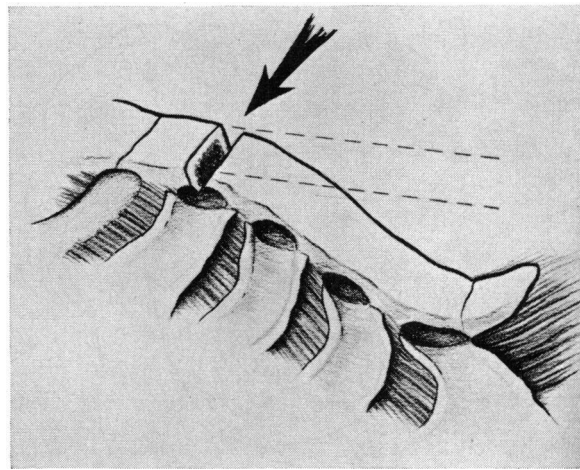


Figure 5.—Cartilages are removed and transverse osteotomy is made permitting straightening of sternum.

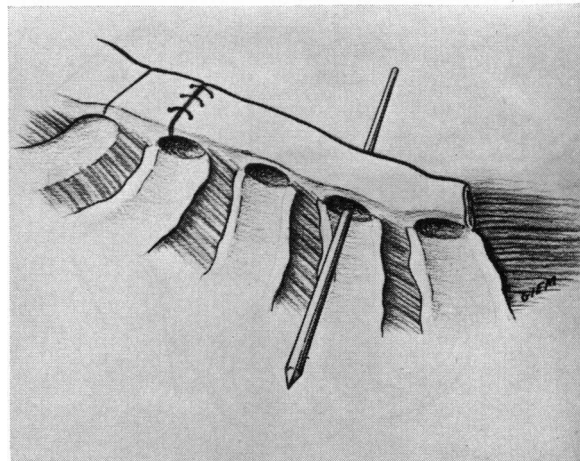


Figure 6.—Sternum is separated from mediastinum and edges of osteotomy are approximated. The Steinmann pin is inserted.

belts or appliances, surgical correction also should be considered.

Some observers advocate "limited" sternoplastic correction in infancy. This procedure had been done previously on one of the patients we treated, and reoperation was required. The optimum age for operation is 3 to 5 years, the results being better both surgically and psychologically.

Surgical Technique

An inverted Y incision is made, with the bifurcation of the incision beginning slightly above the xiphisternal junction. The lateral extent is determined by the deformity. In female patients the lower lateral incisions are contoured to follow the existing or predicted inframammary folds. Keloid formation over the sternum has not been a problem.

The pectoral muscles are elevated with the skin flaps. The perichondrium of each costal cartilage is

incised and stripped free, exposing the cartilage. The cartilage is removed completely, leaving an intact perichondrium. The xiphoid bone is excised by sharp dissection and a wedge transverse osteotomy is made at the upper end of the sternum where the deformity begins. Blunt finger dissection beneath the sternum releases posterior sternal attachments (Figures 4, 5 and 6).

Reconstruction is achieved by reapproximating the rectus muscles to the lowest perichondrial strip. The osteotomy edges are approximated. A Steinmann pin is inserted transversely between the sternal bone plates with each end passing outward through the lateral muscle and skin flaps. This pin provides stability during the fixation phase of healing. The substernal space is drained by a catheter attached to suction.

Usually there are no operative complications. Unilateral pneumothorax, which occurred in three cases, was satisfactorily managed by catheter suction for 24 hours.

Ambulation is begun on the first postoperative day. The pin is removed as soon as the wall of the chest is firm enough to resist a flail motion—usually 10 to 15 days after operation—and exercise to tolerance is allowed by three weeks.

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